

## Two-Dimensional Echocardiography in Double Orifice Mitral Valve

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**In a patient with endocarditis and significant aortic insufficiency, two-dimensional echocardiography revealed an abnormal mitral valve configuration with division of the valve into two separate orifices. At autopsy, a double orifice mitral valve with two sets of valve leaflets was**

**observed. Appreciation of this echocardiographic abnormality is important because double orifice mitral valve is associated with other congenital anomalies and this echocardiographic configuration may be confused with other cardiac abnormalities.**

Double orifice mitral valve is a relatively rare congenital abnormality, usually discovered as an incidental finding at autopsy or during surgical correction of an associated cardiovascular abnormality (1-4). We present the case of a patient in whom echocardiographic evidence of a double orifice mitral valve was obtained before direct visualization of the valve at autopsy.

### Case Report

A 30 year old man presented with a 4 week history of ankle edema and shortness of breath. Four to 5 days before admission, he noted fever and symptoms of an upper respiratory infection. He had a history of a cardiac murmur in childhood, though its origin was unknown.

*Physical examination* revealed a well developed man in no distress. Blood pressure was 134/44 mm Hg. Pulse was 120 beats/min and respirations 22/min. There was mild neck vein distension with hepatojugular reflex. There were dry rales in both lung bases. The first heart sound was loud, as was the pulmonary component of the second heart sound. A third heart sound was audible at the apex. A grade 2/6 systolic ejection murmur heard over the entire precordium was best heard along the left lower sternal border. There was a grade 3/6 long diastolic decrescendo murmur over the left lower sternal border. There was no Austin Flint murmur.

*The electrocardiogram* showed normal sinus rhythm, left

atrial enlargement and nonspecific ST-T wave changes. The chest X-ray film showed cardiomegaly with mild pulmonary vascular congestion. Six blood cultures grew no organisms.

*Two-dimensional echocardiography* performed on admission showed a large vegetation on the aortic valve. The mitral valve had an abnormal configuration (Fig. 1 and 2). The left ventricle was moderately dilated with good contractility. The left atrium was at the upper limits of normal in size. Permanent records of the M-mode echocardiogram could not be obtained because of equipment malfunction.

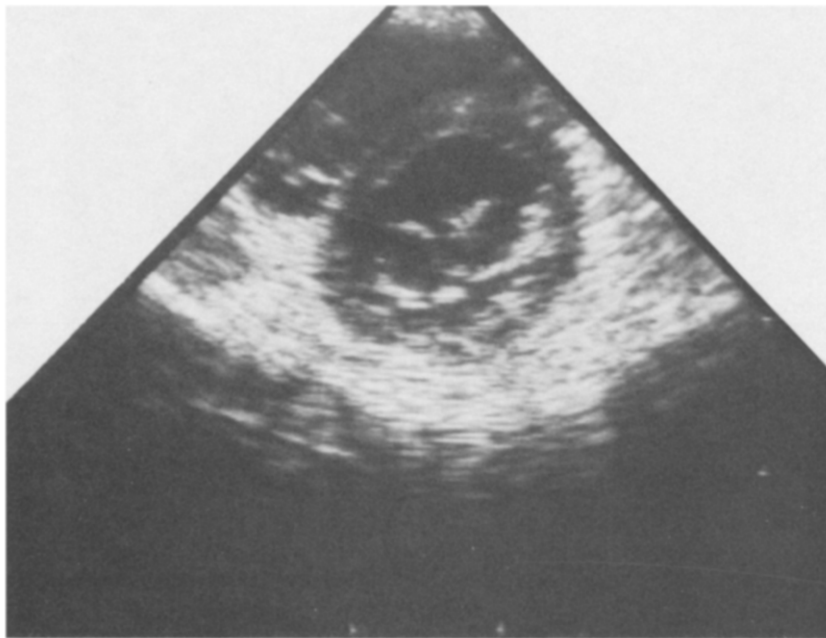
*Cardiac catheterization* revealed a right atrial pressure of 14 mm Hg, mean pulmonary artery wedge pressure of 22 mm Hg and aortic root pressure of 100/50 mm Hg. No attempt was made to cross the aortic valve because of the vegetation. Oxygen saturation determinations showed no evidence of left to right shunt. An aortic root injection showed 4+ aortic insufficiency with good visualization of the left ventricle. The left ventricle was mildly dilated with a mild decrease in contractility. There was no evidence of mitral regurgitation.

Aortic valve replacement was planned for after 4 weeks of intravenous antibiotic therapy. The patient was doing well, having been given diuretic drugs and started on digoxin therapy. Three days before his scheduled operation, the patient developed a sudden episode of tachypnea, the cause of which was uncertain. Chest X-ray examination revealed a right upper lung field infiltration. Blood and sputum cultures failed to yield a causative organism. A Swan-Ganz pulmonary artery catheter showed normal pulmonary artery wedge pressures. The cardiac output was elevated. The patient developed a metabolic acidosis and died within 24 hours.

*At autopsy* the heart weighed 600 g. The right-sided heart structures were normal. The interventricular and interatrial

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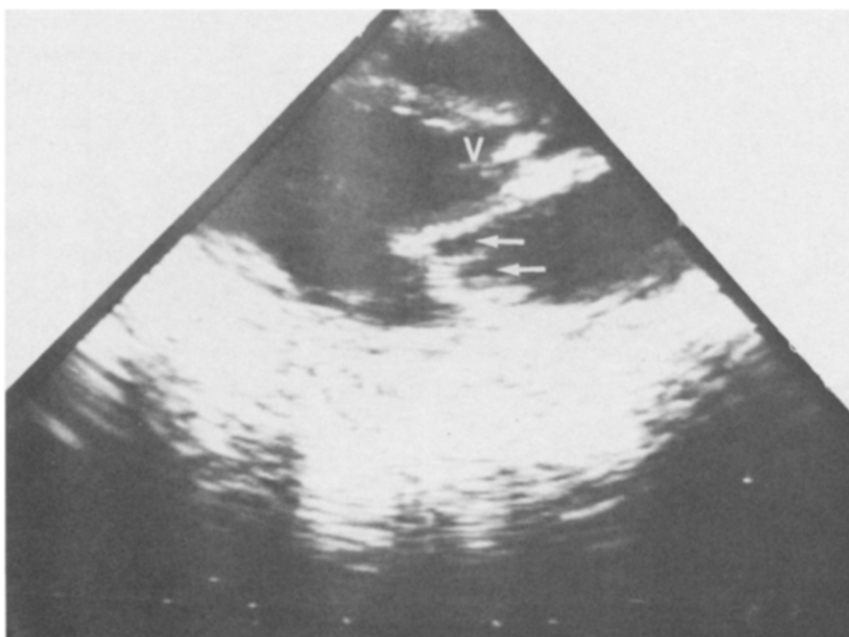
**Figure 1.** Two-dimensional echocardiogram. Parasternal short-axis view at the level of the mitral valve demonstrating two separate mitral orifices of nearly equal size. The anterolateral orifice is to the **right** and the posteromedial orifice is to the **left**.

septa were intact. The mitral valve was composed of two separate valve orifices with separate leaflet structures. The longest diameter of the lumen of the anterolateral valve was 1.7 cm and that of the posteromedial valve was 1.5 cm (Fig. 3). The mitral valve leaflets were smooth without vegetations. The anterior leaflet of the anterolateral valve was connected by chordae tendineae to multiple small papillary muscles that were fused at their tip into a broad band. The remaining leaflets of both valves were connected by chordae tendineae to multiple independent papillary muscles. There was an increase in trabeculation of the left ventricle, with

a few delicate bands bridging the ventricle. Extensive destruction of the aortic valve by endocarditis precluded evaluation of its architecture (Fig. 4). There was no evidence of coarctation of the aorta. A focal interstitial pneumonitis with acute bronchopneumonia was also noted.

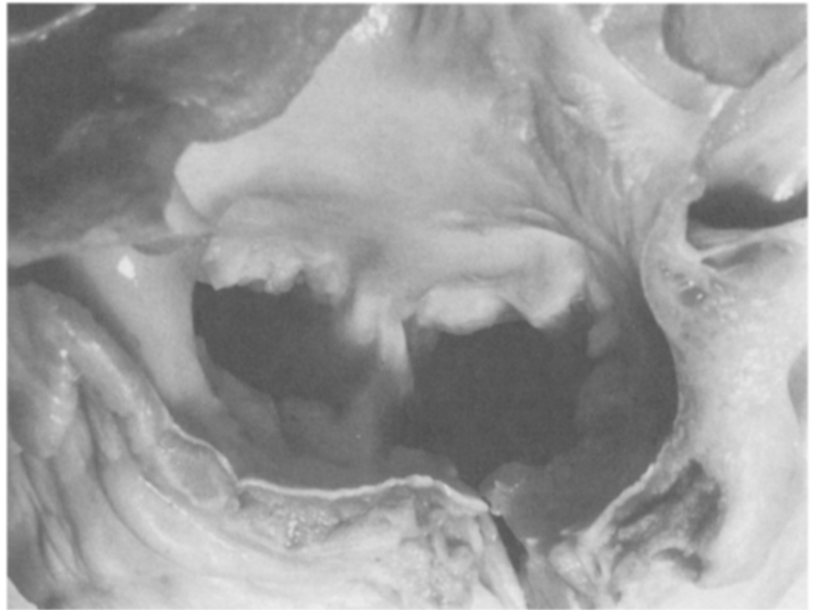
## Discussion

**Association of anomaly with other cardiac abnormalities.** Double orifice mitral valve may occur as an isolated anomaly. More frequently, it is associated with other



**Figure 2.** Two-dimensional echocardiogram. Parasternal long-axis view demonstrating the anterolateral (**upper**) and posteromedial (**lower**) orifices of the mitral valve (**arrows**). Echo-dense vegetation on the aortic valve may also be seen. V = aortic valve vegetation.

**Figure 3.** Autopsy specimen. The two orifices of the mitral valve are seen as viewed from the left atrium. The anterolateral orifice is seen on the **left** and the posteromedial orifice on the **right**.



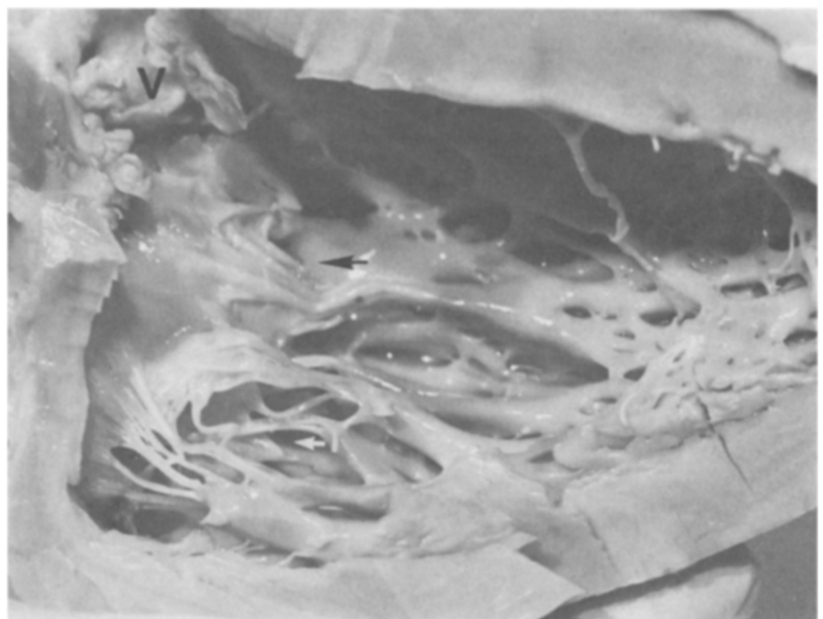
cardiac abnormalities. In descending order of frequency, endocardial cushion defect, bicuspid aortic valve and coarctation of the aorta are the most commonly associated anomalies (5). Patent ductus arteriosus, right-sided aortic arch, subaortic stenosis, bicuspid pulmonary valve, Ebstein's anomaly and secundum atrial septal defect have also been reported (6). In our case, no associated lesions were found, though congenital bicuspid aortic valve could not be excluded because of destruction of the native valve by endocarditis. The absence of associated cardiac abnormalities appears to be more frequent in patients such as ours with relatively equal mitral valve orifices. In their review, Rosenberg and Roberts (5) found that 19 of 30 patients with unequal orifices had associated malformations compared with

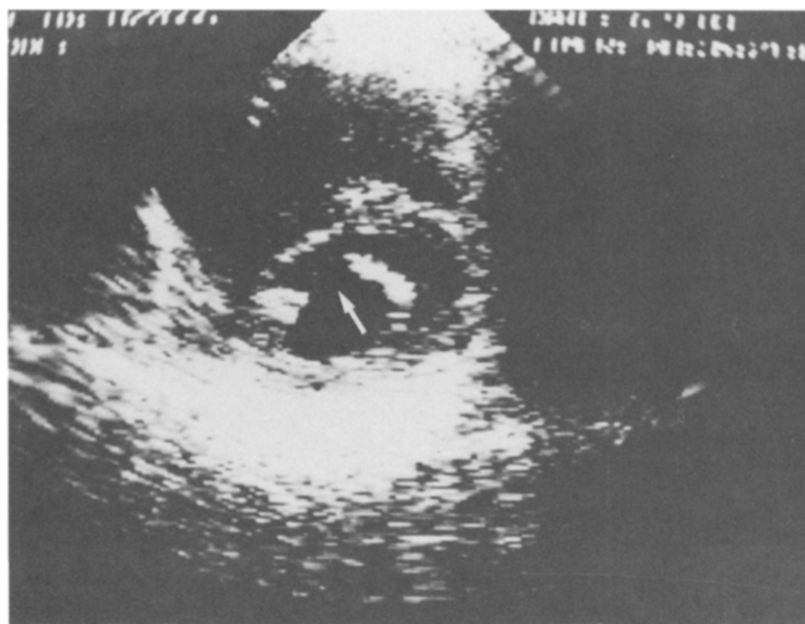
only 2 of 9 patients with equal or nearly equal orifices.

Double orifice mitral valve is usually associated with normal mitral valve function; however, mild to severe mitral regurgitation or significant mitral stenosis, or both, has been reported in some cases (2,6-8). In our case, angiography revealed no evidence of mitral insufficiency. A mitral valve gradient was not measured, but inspection of the valve at autopsy suggested adequate orifice size and mobile leaflets, making significant mitral stenosis unlikely.

**Echocardiographic findings.** The parasternal short-axis view in our patient demonstrated the most obvious echocardiographic findings in double orifice mitral valve (Fig. 1). The two separate valve orifices are clearly demonstrated in this cross-sectional view. The parasternal long-axis view

**Figure 4.** Autopsy specimen. A section through the long axis of the left ventricle shows the anterolateral (**upper**) and posteromedial (**lower**) valves (**arrows**). Note the multiple small papillary muscles, the increased trabeculation of the left ventricle and the vegetations on the aortic valve. V = aortic valve vegetation.





**Figure 5.** Cleft mitral valve. Parasternal short-axis echocardiographic view in early diastole shows defect (**arrow**) in the anterior leaflet of the mitral valve. (Reproduced with permission from the Clayton Foundation for Research, Texas Heart Institute, Houston, Texas.)

likewise demonstrates separate anterior and posterior valve orifices. In the diastolic frame (Fig. 2), at least three separate leaflets perpendicular to the ultrasonic beam can be seen. Apical views were also obtained in this patient and although no single frame showed convincing evidence of the double orifice, real time viewing of the apical four chamber view suggested the presence of the multiple papillary muscles (gross specimen in Fig. 4). Because of an equipment failure, M-mode tracings could not be obtained in our patient. However, others (8) have described some thickening of the mitral valve leaflets on the M-mode echocardiogram and identified two separate mitral valve apparatuses in slightly different echocardiographic planes.

**Differential diagnosis.** Other echocardiographic abnormalities with which double orifice mitral valve might be confused include cleft mitral valve and abnormalities of the anterior leaflet of the mitral valve in aortic insufficiency (Fig. 5 and 6). Because double orifice mitral valve is frequently seen in patients with an endocardial cushion defect, it is important to distinguish it from cleft mitral valve. The cleft mitral valve will appear as a defect in the anterior leaflet of the mitral valve, resulting in a two-sided mitral valve structure but without the complete separation into two orifices as seen in this patient (9). In patients with significant aortic insufficiency, initial diastolic indentation of the anterior mitral valve leaflet by the regurgitant jet may divide



**Figure 6.** Aortic insufficiency. Parasternal short-axis echocardiographic view in early diastole in a patient with severe aortic insufficiency demonstrates indentation of the anterior leaflet of the mitral valve. (Reproduced with permission from Rowe D, et al. and John Wiley & Sons [10].)

the orifice of the mitral valve (10). The anterior leaflet indentation in aortic insufficiency is V-shaped, however, and, therefore, is distinct in appearance from the division between the orifices seen in our patient.

**Implications.** The appreciation of this abnormality may be important, as the lesion is associated with other congenital cardiac anomalies and the echocardiographic configuration may be confused with other cardiac abnormalities.

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## References

1. Bor N, Peters R. Double mitral apparatus or orifice: report of a case. *Arch Pathol* 1957;64:92-9.
2. Yurdakul Y, Bilgic A, Saylam A, Sarioglu T, Kosker S, Aytac A. Congenital double-orifice mitral valve. Report of a case with valve replacement. *Jpn Heart J* 1980;21:545-50.
3. Ancalmo N, Ochsner JL, Mills NL, King TD. Double mitral valve. Report of a case and review of the literature. *Angiology* 1977;28:95-100.
4. Schraft WC, Lisa JR. Duplication of the mitral valve: case report and review of the literature. *Am Heart J* 1950;39:136-40.
5. Rosenberg J, Roberts WC. Double orifice mitral valve: study of the anomaly in two calves and a summary of the literature in humans. *Arch Pathol* 1968;86:77-80.
6. Mercer JL, Tubbs OS. Successful surgical management of double mitral valve with sub-aortic stenosis. *J Thorac Cardiovasc Surg* 1974;67:440-2.
7. Reed GE, Cortes LE, Clauss RH, Reppert EH. The surgical repair of duplication of the mitral orifice. *Ann Thorac Surg* 1970;9:81-5.
8. Warnes C, Somerville J. Double mitral valve orifice in atrioventricular defects. *Br Heart J* 1983;49:59-64.
9. Hagler DJ, Tajik AJ, Seward JB, Mair DD, Ritter DG. Real time wide angle sector echocardiography: atrioventricular canal defects. *Circulation* 1979;59:140-50.
10. Rowe DW, Pechacek LW, DeCastro CM, Garcia E, Hall RJ. Initial diastolic indentation of the mitral valve in aortic insufficiency. *JCU* 1982;10:53-7.